



Original Research Article

Clinical profile, management and outcomes of pseudoexfoliation syndrome with glaucoma in a tertiary eye care hospital

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ABSTRACT

Objective: To study the clinical profile, management and outcomes of pseudoexfoliation (PXF) syndrome with glaucoma in a tertiary eye care hospital.

Materials and Methods: This is a prospective hospital based interventional study over a period of two years from January 2020 to December 2022. 300 eyes of 150 cases aged between forty to ninety years, visiting OPD and diagnosed with PXF syndrome with glaucoma were enrolled in the current study. Cases with other causes of glaucoma, ocular trauma or surgery, cases who were lost on follow-up were excluded.

Results: In our study, 67% eyes with PXF syndrome were non-glaucomatous and 33% (300 eyes) were glaucomatous which was taken as the study population. PXF syndrome cases having glaucoma showed 43 % of PXF glaucoma, 27% primary open angle glaucoma, 18 % primary angle closure glaucoma and 12 % normal tension glaucoma respectively. Bilateral glaucoma was seen in 83% .Mean Intra ocular Pressure (IOP) of the cases was found to be 18.28 ± 3.48 mmHg. Surgical intervention was done in 20% with surgical complications reported among 16.7 % eyes. Mean IOP after 1 month and 12 month of surgery was 16.8 mmHg and 14.6 mmHg respectively. Greater prevalence rate was found with age greater than 60 yrs, mean being 64 yrs and there was no gender predilection.

Conclusions: We observed a greater incidence of PXF syndrome with glaucoma in our region and a positive statistical correlation with increasing age, poor mydriasis and progressive glaucomatous optic neuropathy unresponsive to medical management. Rate of surgical complications was very high. Hence, Ophthalmologists must emphasize on screening and meticulous preoperative examination of PXF to avoid intraoperative complications in these cases.

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1. Introduction

In 1917, Lindberg initially identified Pseudoexfoliation syndrome [PXS] as the deposition of distinctive fibrillar extracellular substance in the anterior segment of eye.¹ Pseudoexfoliation (PXS, PXF, PEX) syndrome is considered as an age related Universal condition with principal ocular exhibition of deposition of whitish grey, fibrogranular amyloid like substance on anterior lens

capsule; zonules; ciliary body; pupillary margin; corneal endothelium etc.^{2,3} About 40 % of PXF syndrome patients develop secondary ocular pathologies, 25% have glaucoma as comorbidity and it is the most recognizable cause of secondary open angle glaucoma globally. Major signs of PXF syndrome include, raised intraocular pressure (IOP); poor mydriasis; zonular weakness; corneal endotheliopathy; iris sphincter transillumination, Sampaolesi's line, pigment deposition in trabecular meshwork, cataract, vitreous liquefaction and greater complications during intra ocular surgery.⁴

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Many cases go undetected because of poor mydriasis obscuring view under slit lamp and due to lack of meticulousness on the part of the examiner.⁴ PXF syndrome eyes frequently exhibit asymptomatic rise in IOP, devoid of any pain or visual disturbance. Thus there is further delay in diagnosis in most of the cases. This in turn, intensifies the threat of PXF syndrome on vision and quality of life in elderly. Prevalence of PXF syndrome fluctuates with age, sex, race, ethnicity etc. In the population above 60 yrs, the prevalence of PXF syndrome is 10 to 20 % globally.⁵ In India, it ranges from 1.8% to 7.4% over the age of 45 years.⁶ When we compare gender-wise, the incidence rate of PXF syndrome is higher among females.⁷ A greater occurrence of narrow and occludable angles with PXF was described from India. Further risk factor for glaucoma is higher grades of angle pigmentation leading to progressive glaucoma in around 50 % of cases.⁸ PEX cases are at greater risk of RNFL thinning as found by OCT.⁹ Cataracts were found commonly among the cases of PXF^{10,11} and it is also associated with zonular dialysis during cataract surgical procedure and post-operative lens displacement.¹² The current research is performed to elucidate the clinical profiles and several difficulties in treatment of eyes with PXF syndrome and glaucoma in a tertiary eye care hospital.

2. Materials and Methods

Prospective hospital based interventional study performed in Ophthalmology department and RIO at SCB medical college and hospital Cuttack, over the period of two years from January 2020 to December 2022. Cases were enrolled in first year and followed up for next one year. 150 cases (300 eyes) visiting the OPD and detected with PXF syndrome with glaucoma, aged between 40 to 90 years were enrolled into the research.

2.1. Inclusion criteria

1. 150 cases (300 eyes) of both sex with age between 40 - 90 yrs with PXF by slit lamp investigation and fulfilling criteria for glaucoma
2. Cases who voluntarily gave the written consent for surgery and research. Helsinki protocol was followed

2.2. Exclusion criteria

1. Cases with developmental, traumatic, secondary glaucoma.
2. Cases who have undergone previous ocular surgery or with history of trauma.
3. Systemic or ocular co-morbidity.
4. Cases with Non-compliance to management and/or regular follow-up of 12 months.

In the present investigation, we have considered glaucoma as IOP greater than 21 mmHg in PEX eyes with

Glaucomatous optic disc having any two of these features: vertical CDR greater than 0.5, asymmetry in CDR greater than 0.2 among 2 eyes, typical thinning of neuro-retinal rim & characteristic wedge shaped nerve fibre layer loss.

All cases were subjected to comprehensive Ophthalmological evaluation - visual acuity assessment, slit-lamp investigation before & after dilation of pupils, Goldmann applanation tonometry, dilated fundus and disc evaluation with +90D lens, gonioscopy, Visual field, Diurnal variation of IOP. Mean IOP was taken from 2 observations of diurnal variation. A variance in mean IOP between PEX eye & non-PEX eye of greater than 4 mmHg was considered clinically significant.

2.3. Statistical analysis

Interpretation was evaluated by students 't' test and P-values ≤ 0.05 was statistically significant.

3. Results

Table 1 shows Mean age among the cases was found to be 63.12 ± 8.22 yrs. Maximum cases were found to be 37% in age between 60-70 years and was statistically significant (p-value 0.0001). 10% of the cases were found to be in age group lesser than 50yrs.

Table 1: Distribution of age

Age (in years)	Number of patients, (%)	P-value
40 to 50	19 (10)	0.0001
51 to 60	27 (18)	
61 to 70	50 (37)	
71 to 80	35 (23)	
Greater than 80	19 (12)	
Total	150 (100)	

Table 2 shows Proportion of males (56 %) was not statistically significant to that of females (44 %) (P-value 0.0007).

Table 2: Gender distribution among the patients

Sex	Frequency (%)	P-value
Male	84 (56)	0.0007
Female	66 (44)	
Total	150	

Table 3 bilaterality of disease among the study population was found to be (83%) and statistically significant (p-value 0.0006).

Table 4 shows - on Gonioscopy, 85% of the eyes had open angles and the remaining 15% had occludable angles out of total 300 eyes (p-value 0.00001).

Mean IOP of the cases was found to be 18.28 ± 3.48 mmHg.

Table 3: Laterality of PXF among the patients

Laterality of Disease	Frequency (%)	P-value
Both Eyes	125 (83)	0.0006
Left Eye	12 (8)	
Right Eye	13 (9)	
Total	150	

Table 4: Gonioscopy of the patients

Gonioscopy	Frequency (%)	P-value
Open angles	230 (85%)	0.00001
Occludable angles	70(15%)	
Total	300	

Table 5 shows Cup-Disc ratio among the eyes(total 300). Cup-Disc ratios of the cases were observed to be 0.4 and 0.5 in 23% of the eyes each. 11% of the eyes could not be evaluated due to hazy media with cataract.

Table 5: Distribution of cup-disc ratio

Cup Disc Ratio	Frequency no, (%)
0.2	16, (5)
0.3	17, (6)
0.4	70, (23)
0.5	70, (23)
0.6	67, (22)
≥0.7	28, (10)
Hazy media	32, (11)
Total	300, (100)

Table 6 shows in our study, 67% eyes with PXF syndrome were non-glaucomatous and 33% which were glaucomatous were taken as the study population i.e 300 eyes. Of the eyes with PXF and glaucoma, 43 % had PXF glaucoma, 27% primary open angle glaucoma, 18 % primary angle closure glaucoma & 12 % normal tension glaucoma correspondingly.

Table 6: Status of the Eyes (N=300) among the patients

Eyes status	Frequency (%)
Pseudoexfoliation Glaucoma (PXG)	129 (43)
Primary Open Angle Glaucoma (POAG)	81 (27)
Primary Angle Closure Glaucoma (PACG)	54 (18)
Normal Tension Glaucoma	36 (12)
Total	300 (100)

Table 7 shows All glaucomatous eyes (n=300) were started with medical management, 20% out of which were subjected to surgical management. Also, no surgical interference has been done for 80% of the total eyes.

Table 8 shows some surgical difficulty was reported in 50 (16.7 %) out of the 58 (20%) eyes which underwent surgery which is significant.

Table 7: Distribution of Surgical Treatment among the patients

Surgical Treatment	Frequency (%)
Yes	58 (20)
No	242 (80)
Total	300(100)

Table 8: Distribution of surgical complication of patients

Surgical complications	(Percentage) N=50 (16.7%)
Intraop complications	
Poor mydriasis	(11%)
Zonular dialysis	(2%)
Runaway rhexis	(1%)
Posterior capsular rent	(1%)
iridodialysis	(1%)
Postop complications	
Anterior uveitis	(6%)
Corneal edema	(7%)
Cystoid macular edema	(4%)
Posterior capsular opacity	(7%)
Decentered IOL	(4%)

4. Discussion

PXF with glaucoma was observed to be more prevalent among older population¹³ and is significant because of the increasing elderly population globally. With our present observation, maximum of the cases was among 60-70 years which was statistically significant (p=0.0001). This was similar to an earlier documented research.¹⁴ The prevalence was found to be very low under the age of 50 years in agreement with overall available documented reports.¹⁵

Chi-square test revealed age and gender differences were not significant. There are contradictory observations of gender distribution in PXF.¹⁶

Cases with PX Glaucoma were observed to exhibit greater IOP with diurnal variations & noticeable spikes that probably caused severe optic neuropathy than cases with primary open angle glaucoma. PX Glaucoma is observed among 25-40% cases with PXF syndrome over time.

We found 67% of eyes with PXF were non-glaucomatous and these were excluded. The observed reports were compared to earlier research done in the regions of South India, where 92% of the research populace was non-glaucomatous.¹⁷ In our observation, the mean IOP of cases was found to be 18.28 ± 3.48 . Elevated IOP of 21mmHg or more & CDR greater than 0.7 were observed among 10% cases. 85% of the eyes exhibited open angle whereas remaining 15% showed occludable angles (p-value 0.00001). Unilateral PXF was seen in 48 - 70% of cases and translated to bilateral disease in up to 50% of cases within 5 - 10 years according to various studies.¹⁸ In our study, most of the cases (83%) showed affection of both eyes followed by right eye (9%) which was statistically significant (p-value 0.0006) 8% of the cases showed disease in left eye.

Mean IOP in the current research population was found to be 18.28 ± 3.48 mmHg. Another cross-sectional research showed 91 cases by Rao et al¹⁹ having 18.00 ± 3.40 as mean IOP in unilateral cases whereas 20.15 ± 1.2 in bilateral PXF patients. Thomson et al²⁰ carried out a cross-sectional research among Southern populace of India where the observed mean IOP was found to be 24 ± 1.4 mmHg.

Out of all glaucomatous eyes which were started with medical management, 6% needed Nd: YAG laser Peripheral Iridotomy. Surgical intervention was done in 20% of the cases, out of which in 8% eyes it was because of failure of medical management. 12% eyes which presented with initial high IOP or advanced disc cupping were operated within 3 days of detection after reducing IOP. Complication due to surgical processes was seen in 16.7% eyes. 8% out of the 20% eyes underwent small incision cataract surgery with rigid PCIOL and trabeculectomy with Mitomycin C (triple procedure). 4% out of the 20% eyes were treated with SICS with rigid PCIOL and surgical peripheral iridotomy followed by topical antiglaucoma medication.

The mean IOP at presentation was 18.28 ± 3.48 mmHg and at the end of 12 months after Peripheral Iridotomy and triple procedure was 14.8 mmHg and 14.4 mm of Hg respectively ($P < 0.001$, ANOVA: Two factor without replication test) which is statistically significant. The mean post-operative IOP after 1 month and 12 months was 16.8 mmHg and 14.6 mmHg respectively.

5. Limitation

This was a hospital OPD based study and does not represent the population. Also, study duration and sample size were less and no long term follow up was conducted. No age matched normal controls were taken for comparison of treatment and outcome. We were not able to reliably evaluate disc damage and visual field in 11% cases with visually significant media opacity due to cataract, where only IOP and gonioscopy findings were considered.

6. Conclusion

We found an increased prevalence of PXF with glaucoma in our region than the national average. A positive statistical correlation of severity of PXF with glaucoma symptoms was seen with - age, poor mydriasis, more severe optic nerve injury at presentation unresponsive to medical management. The response to clinical management was poor and necessitated surgical intervention more than in other forms of glaucoma, having public health implications. Hence, screening of all patients above 40yrs attending OPD, by slit lamp examination to rule out PXF is helpful to start early intervention and for careful preoperative planning, anticipating intra operative problems and postoperative care to prevent IOP spikes to ensure safe operation and effective post-operative outcomes. Hence it is concluded that role of Ophthalmologist in treating PXF does not end with a

successful surgery but requires a continuous lifelong follow-up to rule out development of PXF glaucoma.

7. Conflict of interest

None.

8. Source of Funding

None.

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